A DEFINITE CLINICAL VARIETY OF CEREBRAL ARTERIOSCLEROSIS.*

By Joseph Collins, M.D.,

Sclerosis of the blood-vessels of the brain is sometimes to be found on autopsical examination of individuals who, during life, showed no evidence of the disorder in the skeletal blood-vessels. It occurs as an incident in some organic diseases of the brain, particularly softening and general paresis. It occurs as a sole pathological condition which naturally, if it lasts long enough, will be followed by manifestations of disorder of projection tracts like the motor. When cerebral arteriosclerosis occurs as a primary condition, it causes a pathognomonic clinical picture, upon the existence of which alone the diagnosis is justifiable. It is to the consideration of this clinical picture that I invite your attention.

The symptoms that accompany it are fairly constant, and the clinical picture is not subject to much variation. The subjective symptoms are, indeed, very few, and it is with difficulty oftentimes that they are elicited. The patient complains of fugitive headache, often referred to the occipital region; of slight giddiness often coupled with a sensation of insecurity of station and gait which, however, is not attributed to the giddiness; and of impaired snap or vitality. The headache is usually dull in character, and of variable severity. In some cases it is a conspicuous feature; in others it is not. Not very often is it of the intense agonizing character which is fairly characteristic of arterial hypertonus, such as that occasionally occurring with mitral stenosis and aortic insufficiency. These symptoms may exist for several months, or even years, before other and more striking symptoms call professional attention to the patient. The emotional symptoms, which occasionally are early manifestations, are attacks of meaningless laughter, less often of crying, which do not occur in the spasmodic that is sometimes seen in disseminated sclerosis or in ancient apoplexies, but which are like them without attributable cause and without emotional concomitant. The latter is an important feature; the patient does not feel like laughing or like crying.

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The most striking feature of the disease, however, is the patient's appearance. The individual becomes transformed from

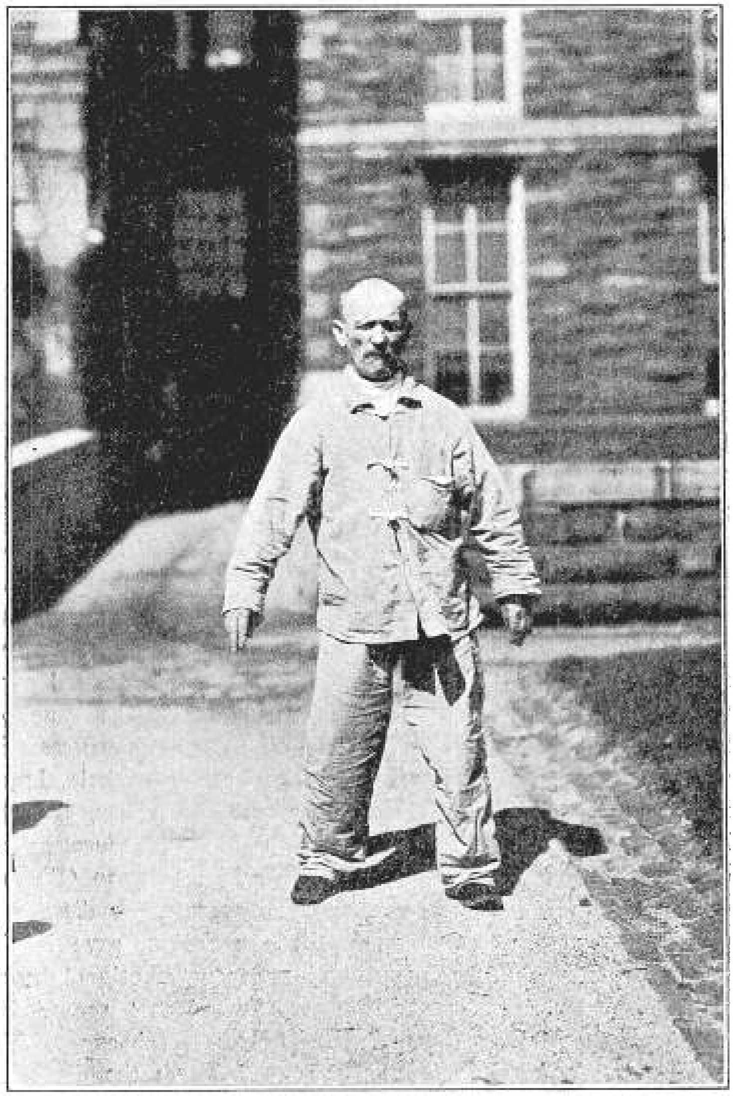

FIG. I.

a person expressing grace in movement and relaxation in repose, into an immobile, inanimate replica of the normal person. The immobilization gives a characteristic attitude and gait, and to a lesser degree, a characteristic physiognomy. The gait is, perhaps, the most remarkable feature of the patient. The stride is short, oftentimes only a few inches, the feet widely separated and not lifted far from the ground, the rhythm of the movement often slow, but sometimes rapid. When the patient turns, he often thrusts out the hands as if to seek support, though he rarely falls. In some instances, perhaps in all, the patient can run better than walk. It is very difficult to show these in a photograph, but in a measure Fig. 1 depicts them. Mental symptoms may or may not exist. In the majority of cases they are not conspicuous, consisting merely of some depression, indifference and apathy. When the disease is far advanced, mental symptoms are more common. At first sight the clinical picture reminds one of Parkinson's disease, but on close observation they have only one feature, immobilization, in common. There are no vasomotor symptoms and secretory symptoms, no marked alteration in the pitch of the voice, no characteristic tremor, no festination, or other striking feature of the latter disease.

The objective symptoms, aside from those that have been enumerated, are few and inconsequential. The knee-jerks are usually lively, and in some cases that have lasted a long time the big toe phenomenon of Babinski is present, indicating secondary degeneration in the pyramidal tracts of the spinal cord, but it is not an essential clinical feature, nor is disturbance of the sphincters, which I have seen in one or two instances. In some cases there is evidence of sclerosis of the skeletal and visceral arteries, but in others, perhaps the majority, there is slight involvement of them. Indeed, the blood-pressure, as revealed by the sphygmomanometer may be low, 110-130 (S) and the heart sounds devoid of particular change. The disease is essentially chronic and subject to little variation, save in the amount of headache, giddiness, and apparent emotional manifestation, which are variable. One of the most remarkable features of the disease is the occasional complete absence of visceral and skeletal arteriosclerosis. In such cases the diagnosis would be difficult because of the absence of increased blood-pressure, of palpatable thickening of the peripheral blood-vessels, and of alteration of the heart sounds, were it not for the fact that the clinical picture which it produces is quite pathognomonic.

In private practice, patients thus afflicted are often looked upon as hysterical or neurasthenic individuals, while in hospitals, especially those given over largely to patients suffering from chronic disease, they go to swell that considerable number that are neither studied nor diagnosticated.

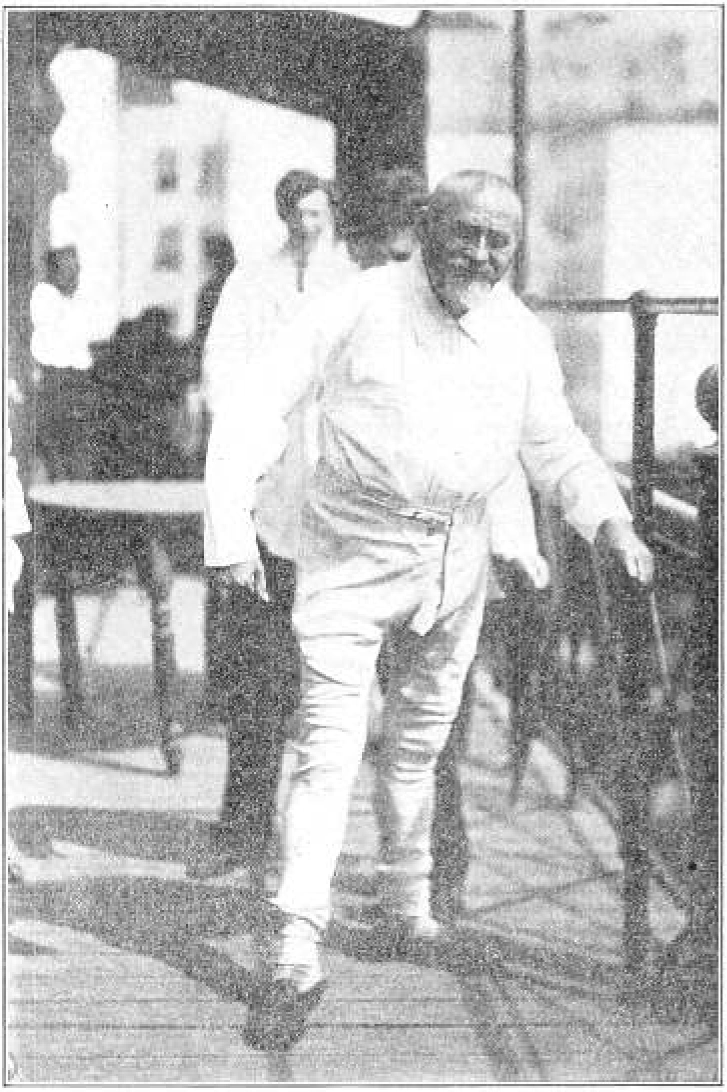

FIG. 2.

The paucity of subjective symptoms and the striking objective symptoms are illustrated by the following history: The patient, a Russian peddler 48 years old, who denies syphilis and alcoholism, noted about six months before he came into the hospital that he was becoming unsteady when he walked and that he often had a sensation of dizziness. At times he had some headache and a buzzing sensation in the head. Further than this there was no complaint. The most remarkable symptoms were his appearance and his gait. The inanimate, wooden transformation was expressed in features and in limbs. When he attempted to walk, the feet, wide apart, shuffled along the floor a few inches at each stride, the strides sometimes fast, sometimes slow, while he made balancing movements with the hands. The only objective symptoms, aside from these apparent to the eye, were those of mild arterial sclerosis. The blood-pressure was 165 (Stanton) ; the vessel walls were palpatable, but there is no hypertrophy of the heart. He has been under observation for a year and very little change has taken place in him during that time, save that the arterial hypertonus has disappeared, as it does in the majority of the cases of arterial sclerosis subjected to hospitalization and iodide of potassium.

Another most typical case of this variety of cerebral arteriosclerosis is the following: The patient, 48 years old, a peddler, whose wife had fifteen children and two miscarriages, began to complain when he was 44 years old of indefinite pains in the legs and of a heaviness in the lower extremities, particularly on going upstairs. About this time his wife noticed that his speech became altered, but she was unable to describe the alteration very lucidly, save to say that it was difficult to understand it. In addition to this, she noticed that he laughed constantly. After these symptoms had lasted for some time, he fell one day as if paralyzed. He did not lose consciousness, but he was paralyzed, —on which side neither he nor his wife knows. This hemiplegia gradually disappeared, and when he got up and began to go about, the peculiar alteration of station and gait which he now has, became evident. The accompanying photograph will give some idea of the station and gait. He stands with the entire body bent forward as if he were a pillar about to topple over. There is no evidence of paralysis. His face is spastic, and he constantly breaks into laughter. When he attempts to walk.

he must hold on to some one; the stride is about two inches, the feet wide apart, shuffle; there is no staggering. After walking about for a while, he gets tired and exhausted.

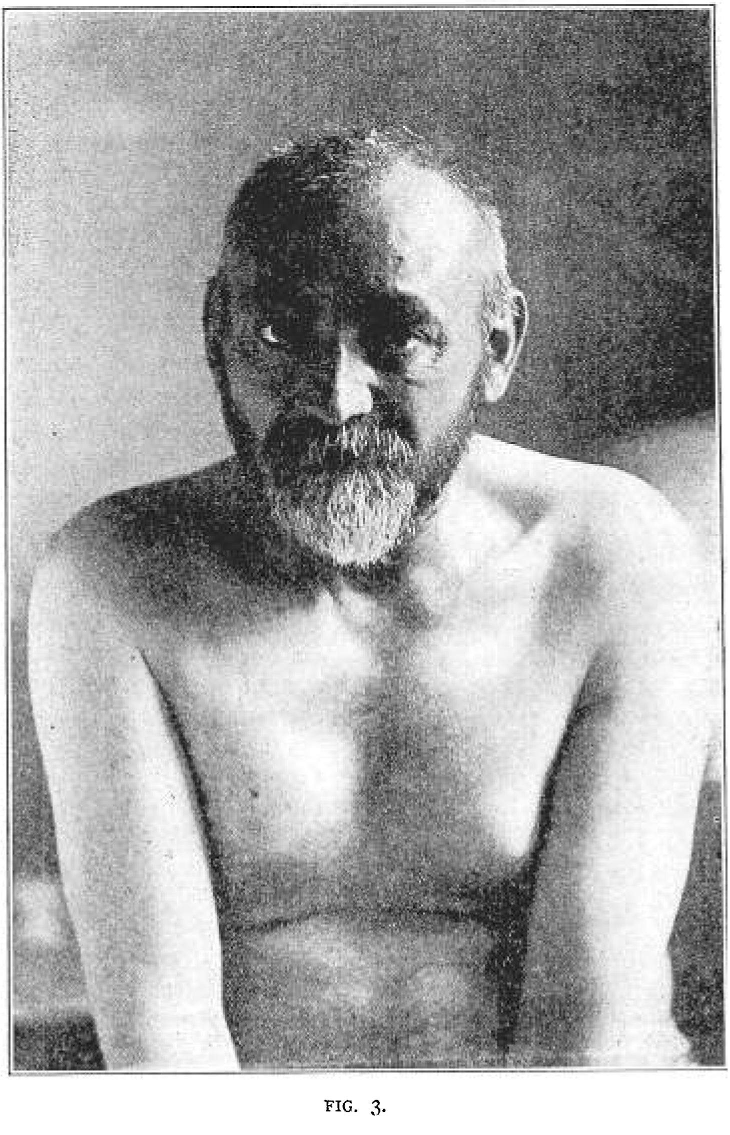

FIG. 3.

Examination shows, in addition to this remarkable immobility which has been mentioned, no particular physical signs save those of the profound arteriosclerosis. There is no evidence of hemiplegia, unless a slight drop of the right angle of the mouth may be so considered. The tendon jerks are not markedly exaggerated. There is no Babinski sign. The superficial reflexes are absent. The speech shows profound dysarthria. He mumbles his words and speaks without particular intonation. There is no difficulty in swallowing, no drooling. It is difficult to estimate his mentality, but he is fully oriented, looks after himself in a normal way, manifests evidences of pleasure when his family visit him, and in a general way comports himself like an individual in possession of his faculties, but whether or not his memory is seriously impaired cannot be said. The condition in which he is now has remained without particular alteration for upward of three years. The manifestations of arteriosclerosis are fairly well marked, but not profound. The blood-pressure is various, from 150-180 (S.) and the radials, temporals, and other palpatable arteries are thickened and resistent. The heart is but slightly hypertrophied, and save for a very common accentuation of the second sound, shows no abnormality. Repeated examinations of the urine shows no albumin or casts save an occasional hyaline cast. Hemoglobin averages 85%, red blood cells 4,000,000, white blood cells 10,000 to 12,000.

These two citations are sufficient, I think, to show the general features of the cases. I am inclined to believe that this variety of arteriosclerosis of the brain will be found to be not extremely uncommon. I have notes of fifteen cases with five autopsies out of a total of 135 cases of cerebral arteriosclerosis. The wonder is that they have not had more attention paid them. In the literature one constantly sees them mentioned to illustrate other points in neuropathology. An excellent example of the disease was presented to the Philadelphia Neurological Society, Nov. 22nd, 1904, by Dr. W. C. Pickett.[1] "A man 65 years old began to have headache and dizziness and became stiff all over, and to have difficulty in walking; mumbling speech and no impairment of mind." Recently Farquhar Buzzard and Stanly Barnes[2] have reported a case of "Chronic Progressive Double Hemiplegia," which is identical with the disease that I am endeavoring here to portray, and which I consider to be a most important and opportune contribution to the recognition and proper interpretation of it. A woman of 43 experienced a trivial fall from which, however, she dated her symptoms, which were attacks of uncontrolable laughter, weakness of the legs and alteration of speech. Ten years later she came under the reporters' observation, her symptoms being then the same as before. "The face was noticeable for some loss of the natural expression. All the facial movements were awkward and slow. Frequent attacks of spasmodic, meaningless laughter. In standing, the back was bowed a little, the head thrown forward. The arms were at the sides, the elbows and hands in partial flexion. She shuffled with small steps, the feet being dragged along sometimes scraping the floor. The knees were bent in walking, but not so much as usual. The whole gait had an aspect of stiffness, and although there was no tremor or propulsion, *yet with her almost expressionless face one was forcibly reminded of paralysis agitans in an early stage.*" Nowhere was any movement completely lost, nor could any difference at any time be made out between the two sides. The tendon-jerks were lively, but not greater than is frequently seen in neurasthenia. The plantar reflexes showed the condition which F. Buzzard has called "pyramidal equilibrium." That is to say, when the plantar jerk was tested with the limb in the general flexed position, the plantar response was invariably flexor, but when tested with the leg and thigh in the extended position, the plantar response was invariably extensor at first, but flexor if the stimulus was repeated. There were no trophic or sensory disturbances. The pulse-rate was 86, medium tension (no sphygmomanometric measurements apparently), no thickening of the vessel wall. The heart and kidneys were apparently normal. She died without warning while eating breakfast. The case apparently had long been considered one of hysteria, but the reporters excluded that chiefly because of the condition of "pyramidal equilibrium" mentioned above, and concluded that the diagnosis was a degeneration occurring in the upper motor neurones, and probably not affecting any other of the main neuronic systems of the central nervous system.

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[1] JOURN. OF NERV. AND MENT. DIS., 1905, p. 182.

[2] Review of Neurology and Psychiatry, 1905, p. 182.

Naked eye and microscopical examination showed most extensive and intense cerebral arteriosclerosis. No gross lesion, such as growth, softening or hemorrhage, was found after hardening in formalin.. The large majority of the arteries, even the smallest, were conspicuous from the rigidity of their walls, and the branches of the perforating arteries on account of a small patch of discoloration which was often present in the tissue surrounding them.

The writers say that "It is remarkable that no sign of the presence of arteriosclerosis could be detected in palpable systemic arteries or in the heart." As I have said in this paper, and frequently elsewhere, this is not uncommon. The writers append a paragraph entitled nomenclature to their article, and favor somewhat, it would appear, the term primary lateral sclerosis, although fully recognizing the inadequacy of such designation. But I venture to suggest, not in a spirit of criticism, that the term chronic progressive double hemiplegia is no more appropriate. Why not call it cerebral arteriosclerosis? That is what it was, and my experience would seem to entitle me to say that in many instances cerebral arteriosclerosis portrays itself in every feature, as shown by the patient of Buzzard and Barnes, and that the picture is absolutely pathognomonic from which the diagnosis can be made even in the absence of every manifestation of general arteriosclerosis.

The gait of these patients, as I have previously said, is one of the most striking features. Naturally this has not escaped the observation of our predecessors. Charcot[3] described an abasia trepidant of the senile, which is not unlike that to which I call attention, if it is not the same thing. Certain it is that the gait of patients with arteriosclerosis, described by the Polish physician, Bieganski,[4] is the condition to which we refer. An article by Petrén,[5] entitled "Ueber den Zusammenhang zwischen anatomisch bedingter und functioneller Gangstörung in Greisenalter," gives an excellent full description of it.

After death, which in some instances comes suddenly from successive attacks of syncope, from intracerebral, intracerebellar, and intraventricular hemorrhage, and from intercurrent trifling disease, such as influenza, acute indigestion, etc., very striking changes are found in the brain.

I have been accustomed to compare the condition to the contracted kidney of arteriosclerosis. Whether there must be a definite localization of the arteriosclerosis to produce this symptom-complex, I am not in a position to state. In the cases that I have with autopsies, the involvement of the vessels has, in the majority of instances, been extensive.

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[3] Leçons du Mardi, 1888-1899, pp. 335, 469.
[4] Medyryna, xiii., 1893.
[5] Archiv für Psychiatrie, xxxiii., xxxiv.

In some instances the entire brain is shrunken; in others it has a normal appearance. The vessels show the essential lesion which varies in distribution, in character, in intensity and in extent. Sometimes, though not always, the vessels at the base of the brain are atheromatous, hard, and gaping when cut across, but in other instances the arteriosclerosis is not apparent until the brain is opened. Then the most striking alterations usually are to be seen in the middle cerebral arteries and their branches.

If death has been immediately conditioned by an accident such as rupture of a blood vessel, naturally the results of this will be found. The alteration of the brain substance depends upon the duration and intensity of the process in the blood vessels, and a detailed description of these to cover every case cannot be given here. In some instances there will be found secondary changes of the nature of Wallerian degeneration in the spinal cord, when the motor tracts of the brain, especially the subcortical motor tracts, are encroached upon.

For instance, a summary of the protocol of the autopsy of the last case that I had may be taken as an example:

1. Small brain.
2. Veins congested.
3. Arteries stand out as thick, cord like structures.
4. Markings typical, but not symmetrical.
5. All the arteries, particularly those of the base, show a very extreme degree of thickening with calcification and fatty degeneration.
6. Areas of nodular dilatation, i. e., aneurysms, practically on all the trunks.
7. Left vertebral artery 1.5 cm. from its origin at the basillar is plugged by a thrombus of very recent origin.
8. The branches of both internal carotids anterior to the circle are much diminished by the thickened walls.
9. Cortical thickness extremely irregular.
10. Macroscopical branches of arteries appear obvious, due to great thickening of walls.
11. Small vessels of pons have a distinct gaping appearance.
12. There is no apparent difference in the degree of arterial change in any portion of the brain.